Parotitis as the Presenting Symptom of Wegener's Granulomatosis: Case Report and Meta-Analysis

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Wegener's granulomatosis is a systemic disease characterized by necrotizing granulomatous lesions and vasculitis of small and medium-sized blood vessels. First reported by Klinger in 1931 [1], the syndrome classically involves the upper airways, lungs and kidneys. Involvement of other organs such as the skin, eyes or the musculoskeletal system is also common [2,3]. Patients often present with severe upper respiratory tract findings such as sinus pain and purulent or bloody nasal discharge. Specific autoantibodies against the neutrophil cytoplasmic enzyme proteinase-3 can be detected and aid in the diagnosis of WG, which is confirmed by the typical granulomatous inflammation seen in biopsies of the affected tissues.

Parotid involvement in WG is unusual and only a few cases have been reported [4-6]. We describe the case of a 57 year old man who developed parotitis, which was the presenting symptom of WG, and review the literature on this unusual presentation and its possible implications.

PATIENT DESCRIPTION

A 57 year old healthy male was admitted to the ear, nose and throat department because of complaints of right ear pain associated with a mucopurulent discharge that lasted a month. He denied fever, night sweats or joint pain. His appetite was good and he was not on any regular medication. He had been treated empirically with antibiotics by his family physician without any improvement.

On admission the patient was stable with a low grade fever. Examination revealed a deviation of the left mandibular angle with a fluctuating mass on the anterior wall of the right external auditory canal and discharge. There was no lymphadenopathy, hepatosplenomegaly, signs of arthritis or rash. His blood tests revealed a normocytic-normochromic anemia (hemoglobin 12.7 g/dl) and leukocytosis of 15,600 cells/µl, with a differential count of 80% neutrophils. He had normal levels of electrolytes, and kidney and liver function were intact. Magnetic resonance imaging revealed right parotid gland enlargement and a deep hypodense lesion without enhancement following gadolinium injection. The differential diagnosis at this stage focused on a tumor versus abscess formation. Ultrasound-guided fine needle aspiration was inconclusive. The right ear mass was drained, and the patient was discharged with the working diagnosis of an infected sebaceous cyst under antibiotic treatment with amoxicillin-clavulanate.

Two weeks later, the patient was readmitted due to the lack of clinical improvement. His examination and blood test results were similar to those of the previous admission, but cultures from the ear discharge revealed Pseudomonas. Acid-fast and fungus stains were negative. A biopsy of the parotid mass revealed a few necrotizing granulomas without evidence of vasculitis. Metronidazole was added to the antibiotic regimen that led to amelioration of his symptoms and the patient was discharged again.

Two weeks later (one month after his first admission) the patient presented with complaints of general weakness, dyspnea, orthopnea, hemoptysis, upper back pain, arthralgia and leg edema. He was admitted to our department. On physical examination he was pale and dyspneic, without tachypnea, and his blood pressure was 150/87. The main physical findings included dullness in the percussion of his lungs, with decreased tactile fremitus, decreased breath sounds, and marked ankle edema. The rest of the systemic examination was unremarkable. Laboratory investigations were significant for anemia (hemoglobin decreased from 12.7 to 7.9 g/dl), leukocytosis (white blood cells 15,080 with 89.2% neutrophils), acute renal failure (urea 27 mg/dl and creatinine 7.95 mg/dl), and erythrocyte sedimentation rate 56 mm/hr. Urine specimen was positive for red blood cells, white blood cells (0–4 per field), nitrites and protein. A 24 hour urine collection yielded 8.7 g/L of protein. Blood and urine cultures were sterile. Chest and neck computed tomography revealed necrotizing parotitis, bilateral nodules in the lungs, and pleural effusion. Kidneys and collecting ducts were normal on renal ultrasonography.

Based on the clinical picture, radiological findings, laboratory results, and the biopsy finding of necrotizing granuloma, the diagnosis of Wegener's granulomatosis was suspected and elevated titers of anti-

WG = Wegener's granulomatosis

PR3 antibodies confirmed the diagnosis. The patient was started on methylprednisone 500 mg/day and cyclophosphamide 50 mg/day with gradual mitigation of symptoms and improvement of his renal function up to normalization.

Many conditions are associated with parotid gland diseases, including: a) bacterial, viral, fungal, chlamydial, mycobacterial and helminthic infections; b) hypersensitivity to organic dusts; c) immune mediated diseases such as rheumatoid arthritis, temporal arteritis, Sjogren's syndrome, and sarcoidosis; and d) malignancies including lymphoma. Parotid involvement in WG is rare and can mimic malignancy, abscess, viral infection or unspecified gland enlargement [3].

Since there is no classic characteristic suggesting parotid involvement, the recognition of WG becomes a diagnostic challenge. In the case described here, the initial presentation was restricted to the parotid gland and the biopsy was inconclusive, leading to a 3 month delay until the diagnosis of WG was reached. The mean time for diagnosing vasculitis is 2 months [6,7], but in certain cases the delay is much longer, up to 20 months. The time for diagnosis in this case is reasonable especially when considering the unusual presentation.

METHODS

With the aim of identifying additional cases of salivary gland involvement in WG and analyzing its impact on disease morbidity and outcome, we performed a MEDLINE (National Library of Medicine, Bethesda, MA) search for additional cases in adult patients reported in the literature. Search terms included "Wegener's granulomatosis" and "parotitis," "parotid gland," "salivary glands," "submandibular gland" or "sublingual gland." The search was last performed in March 2012. Additional reports were obtained by checking the references from the selected studies, case reports and review articles. Publications included in our analysis were case reports of Wegener's granulomatosis involving salivary glands. The initial search algorithm resulted in 65 potential publications of which 36 met our inclusion criteria and were included in our analysis. The following data were extracted from the selected papers: age, gender, symptoms at presentation, additional organ involvement, and outcome.

PR3 = proteinase-3

RESULTS

We found 42 cases (including this case report) of parotid involvement in WG [Table 1]. Epidemiological data were available for 39 patients, 60% men and 40% women, with a mean age of 52 years. The parotid gland was the most common salivary gland involved in WG (78%), followed by the submandibular (36%) and the sublingual gland (2.5%). Only three cases presented with isolated parotid gland involvement [27,28,33]. In most of the cases salivary gland enlargement was accompanied by other organ involvement such as upper respiratory (72%), lower respiratory (61%) and renal (47%). Analysis of the incidence of renal involvement in WG patients with salivary involvement by decades, since 1950, revealed a trend of decrease in renal involvement in the last decades [Figure 1]. A high frequency of neurological manifestations (hearing loss, nerve palsy, etc.) and specifically facial nerve palsy (44% and 17% respectively) were reported among the WG patients with parotid gland involvement. Data on patient survival were available for only 33 patients, and among them 6 deaths occurred (18%).

Among the 41 cases reported in the medical literature, 20 were published in ENT-related journals, 8 in general internal medicine, 9 in rheumatology, 2 in pathology and one in a radiology journal.

DISCUSSION

Parotid gland involvement is a rare manifestation of WG, with a reported incidence of 4% to < 1% [7,31,33]. Among 85 patients with WG described by Fauci et al. [11] in 1983, only one patient had parotid gland involvement. But even though this presentation is rare, several cases of WG associated with salivary gland involvement have been reported [5,9,10,16]. Our MEDLINE search focused on the occurrence of parotid and other salivary gland involvement in WG. Besides our patient, we found 41 additional cases. A mild gender predilection (60% vs. 40% male:female ratio) was found in the patients in our analysis, slightly higher than the ratio reported in the literature [14]. The disease developed in the fourth or fifth decade of life, as described in the literature [39].

The parotid gland was the most common salivary gland involved, followed by the submandibular and the sublingual. Isolated parotid disease as the presenting symptom of WG is extremely rare. Our patient is the fifth patient reported [27,28,33,37], but of five cases three eventually developed upper or lower respiratory tract manifestations, indicating that isolated parotid involvement is rarely the sole manifestation of WG.

Upper respiratory and lower respiratory tract involvement was found in the majority of cases (72% and 61% respectively), but renal involvement was described in only 47%, rates lower than those described by Hoffman et al. [7] for 158 patients with WG, where 93% had upper respiratory involvement, 66% pulmonary involvement, and 77% renal failure [7]. Although the classic triad of WG (upper respiratory symptoms, lower respiratory symptoms, kidney vasculitis) is not always present at the time of the initial evaluation, approxi-

Figure 1. Renal involvement in WG by decades

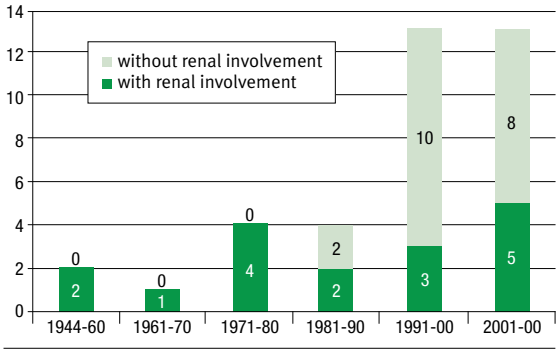

Table 1. Wegener's granulomatosis with parotid involvement

Case [Ref]	Journal	Year of publication	Age/ gender	Salivary gland involvement			Presenting symptom	Renal disease	Other organ involvement'	Outcome
				Parotid	Submandibular	Sublingual				
Lindsay et al.*[8]	*Am J Pathol*	1944	67/F	+			ENT, lung	+	Parotitis (unilateral), eye, heart, pituitary	Died
Fahey et al. [9]	*Am J Med*	1954	39/M	+			Parotid (bil), ENT, eye, orchitis, ulnar neuritis	+	Lung, cardiac	Died
Berman et al. [1]	*Ann Intern Med*	1963	34/M	+			Parotid & submaxillary (bil), ENT, eye	+	Pericard	Died
Fauci & Wolff [10]	*Medicine*	1973	40/M	+			Parotid, ENT, lung, skin, hearing loss	+		Remission
Smith & Konrad [11]	*Arch Otolaryngol*	1976	53/F		+		Submandibular (lt), ENT, lung, nerve palsy	+		Remission
Kovarsky [12]	*Arthitis Rheum*	1978	43/M	+			Lung, ENT, arthralgia, parotitis (rt)	+		Remission
Small et al. [13]	*J Rheumatol*	1980	59/M		+		Submandibular (bil), arthralgia	+	Lung, ENT	Remission
Fauci et al. [14]	*Ann Intern Med*	1983	M	+			Parotid	No information		
Bachmayer et al. [15]	*Wien Klin Wochnschr*	1984	60/F		+		Submandibular, facialis, eye, GI, ENT	+	Liver, pancreas, lung	
Kavanaugh & Huston [16]	*Am J Med*	1988	75/M	+			Parotid (lt), ENT	–	Lung, eye (lt), neurology (6th nerve palsy)	No information
Devaney et al. [17]	*Am J Surg Pathol*	1990	No information	+	+		No information			
Murty et al. [18]	*J Laryngol Otol*	1990	23/M		+		ENT, arthralgia, submandibular (lt)	–	–	Remission
			55/F	+			Parotid mass (lt), ENT	+		
Specks et al. [19]	*Arch Otol Head Neck Surg*	1991	60/M		+		Submandibular (bil), lung, ENT, hearing loss (bil), prostate	–		Remission
			64/M		+		ENT, submandibular (rt)	–	Scleritis	Remission
			24/F		+		ENT, submandibular (rt)	–	Lung	Remission
			27/M	+			ENT, parotid (rt)	–	–	Remission
			75/F	+			Parotid (bil), lung	–		Remission
Stuckey & Smart [20]	*Australas Radiol*	1992	45/M	+	+		Pancreatitis, hematuria (non-glomerular)	–	Parotitis (bil), submandibular (bil), lung	Remission
Benson-Mitchell et al. [21]	*J Laryngol Otol*	1993	34/F	+			Parotitis (unilateral) and hearing loss	–		Remission
Vanhauwaert et al. [22]	*Post Grad Med*	1993	60/F		+		Submandibular (bil), ENT, eye, pancreatitis	+		
Lustman et al. [23]	*Oral Surg Oral Med Pathol*	1994	40/F	+			Lung	–	Parotitis (bil), hearing loss, arthralgia, ENT	Remission
Ah-see et al. [5]	*J Laryngol Otol*	1996	48/F	+	+		Parotitis & submandibular (bil), ENT	–	–	Remission
Ross et al. [24]	*Br J Rheumatol*	1996	66/F	+			Parotitis (bil), ENT, hearing loss, lung	+		Died
Singh et al. [25]	*Sacoidosis Vasc Diff Lung Dis*	1997		+			Parotitis	Not available		
Berge et al. [26]	*Int J Oral Maxillofac Surg*	2000	47/F	+	+		Parotitis & submandibular(rt), Pericarditis, facial palsy (rt)	–	Lung	Remission

Case [Ref]	Journal	Year of publication	Age/ gender	Salivary gland involvement			Presenting symptom	Renal disease	Other organ involvement'	Outcome
				Parotid	Submandibular	Sublingual				
Saravanappa et al. [27]	*J Otolaryngol*	2000	69/M	+			Parotitis (rt)	+	Arthralgia, diarrhea, rash, facial palsy	Remission
Garcia-Porrua et al. [28]	*Rheumatology*	2001	60/M		+		Submandibular (lt), dysphagia	–	–	Remission
Crean SJ et al. [29]	*Int J Oral Maxillofac Surg*	2002	55/M			+	Sublingual (lt), ENT	+		Remission
Liu et al. [30]	*J Laryngol Otol*	2003	46/F	+	+		Parotid & submandibular (bil), lung, ENT	+		Remission
Imamoglu et al. [31]	*Otolaryngol Head Neck Surg*	2003	45/M	+			Parotitis (lt), facial palsy, lung	–		Died
Bulbul et al. [32]	*Med Princ Prac*	2003	45/M	+			Parotitis (lt), lung, facial palsy	–		
Chegar & Kelley [33]	*Laryngoscope*	2004	55/M	+			Parotitis (rt),	+	ENT, facial palsy (rt), lung	Remission
Jones et al. [4]	*J Laryngol Otol*	2005	59/M	+			Scleritis, parotitis	–	Eye, ENT	Remission
Heintz et al. [34]	*Sarcoidosis Vasc Diff Lung Dis*	2005		+			Parotitis	Not available		
Franz et al. [3]	*Eur Arch Otorhinolaryngol*	2006	71/F	+			Parotitis (lt), lung	–		Remission
Ullrich et al. [35]	*Otolaryngology*	2006	60/M	+			Parotitis (rt), ENT	–		Remission
Danda et al. [6]	*Clin Rheumatol*	2008	50/F	+			Parotitis (bil), arthritis	+	Neurology, lung, eye	Remission
Yamamuto et al. [36]	*J Rheumatol*	2008	62/M	+			Arthralgia, neuropathy, lung	–	Neurology, ENT, parotitis (bil), nerve palsy (bil)	Remission
Green et al. [present case]		2009	57/M	+			Parotitis	+	Lung, ENT, arthralgia	Remission
Gassling et al. [37]	*J Rheumatol*	2010	68/M	+	+		Parotitis	+	ENT, eye	Died
Barrett et al. [38]	*Br J Oral Maxillofac Surg*	2011	47/M	+			Parotitis	+	Lung, eye	Chronic renal failure

*Although the diagnosis was periarteritis nodosa, on reviewing the report it seems that the patient had WG

ENT = ear, nose and throat involvement, Lung = lung involvement including hemoptysis and chest X-ray abnormality, M = male, F = female, bil = bilateral, lt = left, rt = right, GI = gastrointestinal

mately 90% of patients with typical WG go on to develop renal disease [3]. The lower incidence of renal involvement in our analysis suggests that these patients may be at a lower risk for development of renal disease. There may be several explanations for this finding: a) parotid involvement belongs more to a local or restricted disease phenotype than to a systemic one, b) parotid involvement may be an early feature of the disease which probably explains the sparing of the kidney on presentation [26], and c) since the parotid gland is a superficial and easily visible organ, it may serve as a marker for early disease evaluation and is more accessible for biopsy (with lower morbidity as compared to lung or kidney biopsy), hence promoting earlier diagnosis and treatment and preventing disease progression. Specks et al. [19] also recognized the lower morbidity and mortality associated with parotid involvement at presentation of WG and assumed that the earlier the patient receives treatment the lesser the likelihood of developing irreversible loss of organ function (early diagnosis and treatment may prevent the progression to systemic disease). In agreement with these speculations, our analysis of the incidence of renal involvement by decades revealed a trend of decrease in renal involvement in recent decades, probably reflecting earlier diagnosis and treatment [Figure 1].

On the other hand, biopsies from extrapulmonary sites, such as the parotid gland, frequently show only non-specific inflammatory changes (as in our patient), and often an open lung biopsy is required to reach a definite diagnosis [16,17]. Devaney and co-authors [17] found that vasculitis, necrosis and granulomatosis were present in only 16% of all head and neck biopsies. Furthermore, interpretation of head and neck biopsies has a wide differential diagnosis, making the diagnosis even more

difficult [17]. Lustman et al. [23] reported similar results, with only 30% of parotid gland biopsies showing the presence of vasculitis.

An increased frequency of neuropathy, especially facial nerve palsy, was reported in the patients with parotid disease as compared to patients with typical WG (41% vs. 22%) [14]. The facial nerve involved was on the same side as the involved salivary gland in all cases that the side was mentioned, pointing to an anatomic connection between the two symptoms. A similar observation was reported by Yamamoto and team [36], who found facial neuropathy in 35% of patients with WG and parotitis.

Interestingly, half of all cases with WG and parotid involvement were reported in ENT journals, and one-quarter in internal medicine and in rheumatology journals each, illustrating the lack of awareness of this unusual presentation.

Although WG is considered an autoimmune disease, several authors have suggested a role for infection and specifically for *S. aureus* in disease development and in relapses [40]. Since *S. aureus* is an important etiological cause of parotid gland infections, it has been suggested that *S. aureus* phosphatase acts as an antigen and an initiator of autoimmune responses leading to vasculitis and glumerulonephritis [25]. The parotid involvement in WG reinforces the possible role of infections and specifically of *S. aureus* in this disease.

In summary, we present a patient and an analysis of an additional 41 cases of WG WG with parotid involvement. Our analysis demonstrates that parotitis can be the initial symptom of systemic WG and should be considered in the differential diagnosis of parotid disease. As such, titers of C-anti neutrophil cytoplasmic antibodies and anti-PR3 antibodies should be measured during the evaluation of parotid enlargement since early treatment and diagnosis may lower the risk for renal involvement.

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